DATE TABLE

TABLE I: LISTING OF SPONGIFORM ENCEPHALOPATHIES (PRION DISEASES) from Prusiner, 1998; Will, 1999, & Taylor, 2002

DISEASE	HOST	MECHANISM OF PATHOGENESIS
Scrapie	Sheep, goat, mouflon	Infection in genetically infectable
Transmissible Mink Encephalopathy (TME)	Mink	Infection from sheep or cattle
Chronic Wasting Disease (CWD)	Mule deer, elk	Spontaneous mutation or unknown
Feline spongiform encephalopathy (FSE)	Cats, Cheetah, puma, ocelot	Infection with prion-contaminated bovine tissues or comtaminated bone meal.
Exotic Ungulate encephalopathy	Greater kudu, nyala, oryx	Infection with prion-contaminated bone meal.
Kuru	Human	Infection through ritualistic cannabalism.
Iatrogenic Creuzfeldt-Jakob Disease (iCJD)	Human	Infection from prion-condtaminated Human Growth Hormone, dura mater, grafts, contaminated surgical instruments.
Creutzfeldt-Jakob Disease (CJD)	Human	Infection from spontaneous mutation
Variant Creuzfeldt-Jakob Disease nvCJD)	Human	Infection from bovine, deer, or elk prions.
Familial Creuzfeldt-Jacob Disease (fCJD)	Human	Germ-line muttions in PrP gene.
Gerstmann-Straussler-Sheinker Disease (GSS)	Human	Germ-line mutation in PrP gene.
Fatal Familial Insomnia (FFI)	Human	Somatic mutation or spontaneous conversion of PrP^{c} into PrP^{Sc}.
Fatal Sporatic Familial Insomnia (sCJD)	Human	Somatic mutation or spontaneous conversion of PrP^{c} into PrP^{Sc}.

TABLE II: PRION DISTRIBUTION IN TISSUES by Ramasamy et al. 2003

TABLE A: PrPC distribution in non-neuronal tissues

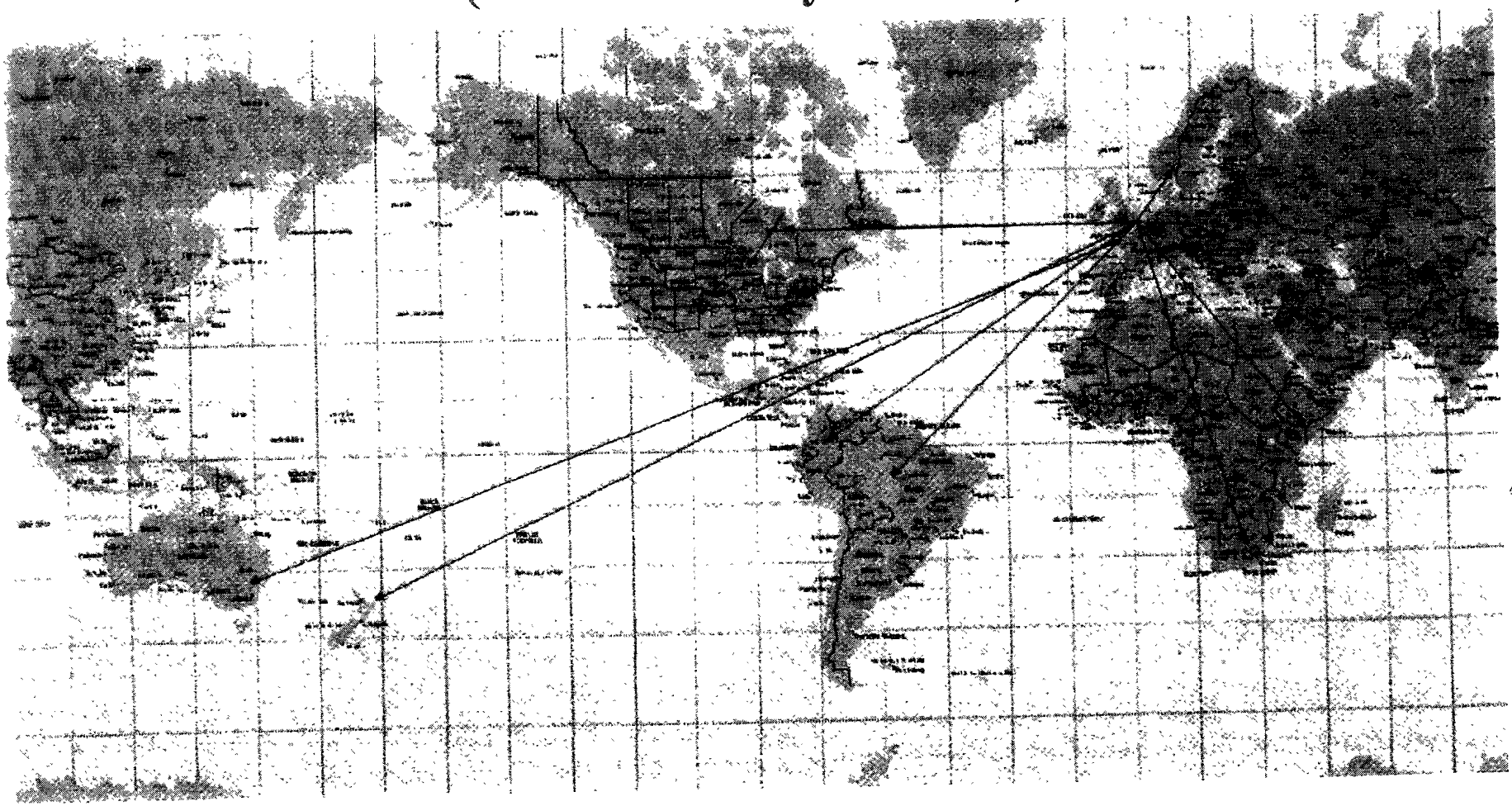
Tissue	Species	Prion strain	Detection method
Cerebral tissues			
Glial cells—eg, astrocytes	Hamsters and rats	PrP^c	In-situ hybridisation[44]
Circulating blood			
B and T lymphocytes and monocytes	Human beings	PrP^c	Immunoblot and cytofluorometry[45,46]
Platelets	Human beings	PrP^c	Immunoblot, timeresolved dissociation-enhanced immunoassay[47,48,49]
Lymphoreticular system Follicular dendritic cell	Mouse	PrP^c	Immunocytochemistry[50,51,52]
Gastrointestinal tract			
Parietal cells	Hamster	PrP^c	Immunofluorescence[53]
Mucous epithelial cells	Human beings	PrP^c	Anti-PrP antibodies[54]
Epithelial cells of the GIT	Human beings	PrP^c	Not stated[55]
Skin			
Keratinocytes in basal layer (cell culture)	Human beings	PrP^c	Western blot[56]
Muscle			
Neuromuscular junction	Human beings	PrP^c	Immunohistofluorescence[57]
Skeletal muscle	Hamster, sheep, mice, cattle		Western blot[52,58,59]
Testes			
Sperm cells	Human beings	PrP^c	Immunoblot detected a truncated prion protein[60]

TABLE B: Tissue distribution PrP^{sc} in people*

Tissue	Prion protein concentration relative to brain	Approximate prion concentration ID_{50}/g tissue*	Comments	Reference
Tonsil	5–15%	10^7	Technique used: western blotting	**Wadsworth et al. (2001)**
Spinal cord	30%	10^7	Technique used: western blotting	
Lymph node	0·1–1%	10^5–10^6	Technique used: western blotting	
Rectum, thymus, and adrenal gland	1/50 000	10^3	Seen in a single patient with high brain prion titres. Technique used: western blotting	
Spleen		10^6–10^7	Technique used: western blotting and bioassay	**Bruce et al. (2001) & Vitale et al. (2001)**

*Based on prion concentrations in the brain of 10^8 ID_{50}/g tissue. Adapted from **Wadsworth et al. (2001).**

TABLE III: SPREAD OF SCRAPIE FROM THE UNITED KINGDOM TO OTHER AREAS OF THE WORLD, 1938-1977 (Detwiler & Bayliss 2003)



Australia	:	1952
Brazil	:	1977
Canada	:	1938
Colombia	:	1968-1971
Kenya	:	1970
New Zealand	:	1952-1954
Norway	:	1958-1959
South Africa	:	1964-1972
United States of America	:	1947 (through Canada)

TABLE IV: Relative Infectivity of Tissues From Sheep and Goat Infected With Scrapie (Detwiler & Baylis 2003)

Table A: Relative Infectivity of Tissues from Suffolk Sheep Naturally Infected with Scrapie

Relative Infectivity	Tissues	References
Highest Levels	Brain, Spinal Cord	Hadlow et al. 1982
Moderate Levels	Lymph Nodes (retropharyngeal, mesenteric-portal, prescapular, pre-temporal, etc.) spleen, tonsil, ileum, proximal colon.	Hadlow et al.1982
	Peripheral nerves (N. ischiadicus, N. axillaries, N. ulnarius, N. medianus, N. tibialis, N. fibularis)	Groschup et al. 1996
Low Levels	Cerebral Spinal Fluid (CSF), sciatic nerve, pituitary gland, nasal mucosa, adrenal gland, distal colon, pancreas, liver, bone marrow, thymus, supra-mammary lymph node.	Hadlow et al. 1982
	N. saphenus	Groschup et al. 1996
No Detectable Infectivity	Blood clot, mandibular and parotid salivary glands, thyroid, heart, lung, kidney, skeletal muscle, mammary gland, testis.	Hadlow et al. 1982

It is important to note that the above levels of infectivity reflect titres at the clinical stage of disease. In the preclinical stage of the disease the titres in the lymphoreticular tissue are actually higher than those in the central nervous system.

Table B. Relative Infectivity from Goats Experimentally Infected with Scrapie

(intracerebral and subcutaneous inoculation)

Relative Infectivity	**Tissues**	**Reference**
Highest Levels	Brain*, Spinal Cord	Hadlow et al. 1974
Moderate Levels	Retropharyngeral, pre-scapular, pre-frontal lymph nodes, spleen, tonsil, adrenal gland*.	Hadlow et al. 1974
Low Levels	Cerebral Spinal Fluid (CSF), sciatic nerve*, pituitary gland*, nasal mucosa, ileum, proximal colon, distal colon, liver, thymus, mediastinal-bronchial lymph nodes, mesenteric-portal lymph nodes, parotid salivary gland*.	Hadllow et al. 1974
No Detectable Infectivioty	Blood clot, submaxillary salivary gland, thyroid, heart, lung, kidney, skeletal muscle, bone marrow, pancreas, ovary, saliva.	Hadlow et al. 1974

*Pattison and Milson (1962) also detected infectivity.

TABLE V: CASES OF BSE AND nvCJD IN VARIOUS COUNTRIES from Will (1999)

COUNTRY	CASES OF BSE	CASES OF nvCJD
United Kingdom	176,425	42
Ireland	345	0
Switzerland	282	0
Portugal	195	0
France	50	1
Belgium	7	0
Germany	6	0
The Netherlands	4	0
Liechtenstein	2	0
Oman	2	0
Italy	2	0
Luxembourg	1	0
Canada	1	0
Denmark	1	0
Falkland Islands	1	0
United States	0	0

TABLE VI: CASES OF INDIGENOUS BSE RECORDED BETWEEN 1994 AND 2001 by Taylor (2002)

COUNTRY	1994	1995	1996	1997	1998	1999	2000	2001
Great Britain	23945	14302	8016	4312	3179	2133	1311	311
N. Ireland	345	173	74	23	18	7	14	6
Belgium	0	0	0	1	6	3	9	19
Slovakia	0	0	0	0	0	0	0	1
Denmark	0	0	0	0	0	0	1	2
France	4	3	12	6	18	31	161	69
Germany	0	0	0	0	0	0	7	90
Greece	0	0	0	0	0	0	0	0
Irish Republic	18	15	73	80	83	91	149	56
Lichtenstein	0	0	0	0	2	0	-	-
Luxembourg	0	0	0	0	0	0	0	0
TheNetherlands	0	0	0	2	2	2	2	11
Portugal	12	14	29	30	106	170	163	44
Spain	0	0	0	0	0	0	2	2
Switzerland	64	68	45	38	14	50	54	19

TABLE VII: DISTRIBUTION OF CASES OF VARIANT CREUTZFELDT-JAKOB DISEASE IN THE UNITED KINGDOM[a] UP TO JANUARY 2003 (P. G. Smith (2003)

Year Person Died	Number of Cases
1995	3
1996	10
1997	10
1998	18
1999	15
2000	28
2001	20
2002	17
Total Deaths	121[b]
Cases Alive	8
Total Cases	129

a: Cases outside the United Kingdom: 6 in France, 1 in Ireland, and 1 in Italy. There has also been 1 case in the United States and 1 case in Canada of people who had previously lived in the United Kingdom in the 1980's and early 1990's.

b: Includes 27 cases without neuropathological confirmation.

TABLE VII: DISTRIBUTION OF CASES OF VARIANT CREUTZFELDT-JAKOB DISEASE IN THE UNITED KINGDOM[a] UP TO JANUARY 2003 (P. G. Smith (2003)

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